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Diffuse hemangioma of the rectum detected on multi-slice CT in an 18-year-old woman with Klippel-Trenaunay syndrome

Klippel-Trenaunay syndrome (KTS) is a rare congenital disorder related to vascular malformations. It is characterized by cutaneous hemangiomas, varicosity and soft tissue and/or bony hypertrophy of extremities. In association with arteriovenous fistula, it is called Klippel-Trenaunay-Parkes-Weber syndrome (3). In about 75% of cases manifestation occurs when the patient is less than 10 years old (2, 10, 11). In very rare cases patients with KTS may develop visceral hemangiomas and thombocytopenia (5). The aim of the present paper is to present a case of colorectal diffuse hemangioma detected on multi-slice CT in an 18-year-old woman with KTS diagnosed in her childhood.

CASE REPORT

An 18-year-old woman with KTS diagnosed in childhood was admitted to the Department of Urology because of a painless hematuria episode, which occurred for the first time; moreover the patient has never had any symptoms of bleeding from the lower alimentary tract. On admission the patient did not complain of any subjective symptoms, except for general malaise. US examination of the abdominal cavity showed no pathological lesions of the urinary tracts. Physical examination presented typical symptoms: hypertrophy of the right lower limb, varices and cutaneous hemangiomas in several remote locations. Laboratory tests revealed the features of mild anemia, clotting disturbances and decreased fibrinogen levels in plasma.

Cystoscopy was performed to assess the bleeding site location. It showed normal urinary bladder volume and mucosa. The left ureteric orifice open wide into the bladder contained normal urine. The right orifice was wide, raised, and contained urine and blood. Following the cystoscopy, it was decided to perform abdominal and pelvic contrast-enhanced computed tomography.

CT was conducted using 8-row computed tomograph Light Speed Ultra (GE Milwaukee Wis) with workstation Advantage Window 4.0. 500 ml of water were used as an oral contrast medium. Due to clinical indications, the examination covered the abdominal cavity, pelvis and the region of proximal thighs. Pre-contrast scanning with collimation 2.5 mm and three-phase post-contrast scanning with collimation 1.25 mm were performed. A non-ionic contrast medium (100 ml Ultravist 300, Schering) was administered with a power injector with the speed of 4 ml/s and the delay time was determined using the Smart Prep technique with density measurements conducted in the abdominal aorta lumen. The delay for the arterial phase was 25 s, for the parenchymatous phase-70 s and for the excretory one-6 min.

The original scans obtained were analysed using multiplanar reformations with maximum intensity mode (MIP) and 3D volume rendering for the evaluation of vascular structures and virtual cystoscopy imaging. MSCT showed enlarged liver and spleen with few, low-echogenic, round foci, up to 15 mm

in diameter, portal hypertension symptoms: wide portal, splenic, superior and inferior mesenteric veins and dilated pericholecystic plexi (Fig.1) The abdominal arteries, particularly those supplying the pelvic organs showed typical branches and diameters. The renal size as well as the excretory function demonstrated no abnormalities, however, the right ureter showed partial contrast enhancement, the width of both ureters was normal. In the central lower pole of the right kidney an uncharacteristic, hypodense, irregular, 2-cm lesion was visible in the parenchymatous and excretory phases, which was situated calyceally (Fig. 2). The bladder was only partially filled, yet no thickening of its wall or indentation lesions were observed. The dilated venous structures of the integuments of the right abdominal wall, gluteal and right iliofemoral regions with hypertrophy of soft tissues were typical of KTS (Fig. 3a, b). In the pelvis, single phleboliths were visible. It was observed that the wall of the rectum and sigmoid was circularly thickened to 2–3 cm, however, the pathological part of the wall contained few phleboliths indicating the vascular character of the tumour (Fig. 4a-c). In the arterial phase the density of the abnormal wall was relatively homogenous being 40 HU and reached 70 HU in the parenchymatous phase. In the excretory phase the density of hemangioma was maintained at the level of 50–60HU. The lumen of the pathological colon was constricted, the contour smooth without features of infiltration of the surrounding fatty tissue. Due to characteristic, hemangioma-like intestinal tumour picture, lack of clinical signs and risk of bleeding on colonoscopy or barium enema, the patient was qualified for conservative treatment and MRI was planned to be made after several weeks.

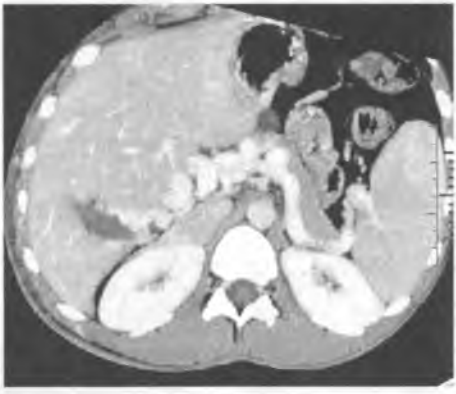

Fig. 1. Symptoms of portal hypertension in oblique MIP reconstruction

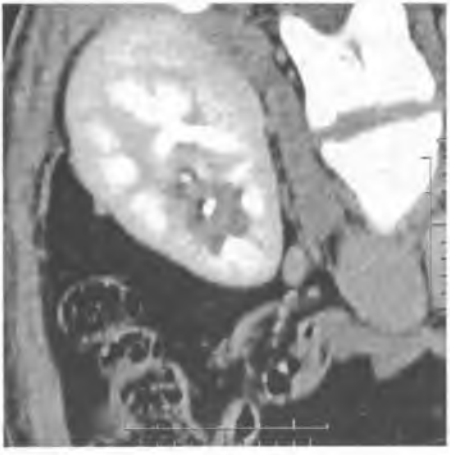

Fig. 2. An atypical lesion in the lower pole of the right kidney in the parenchymatous phase

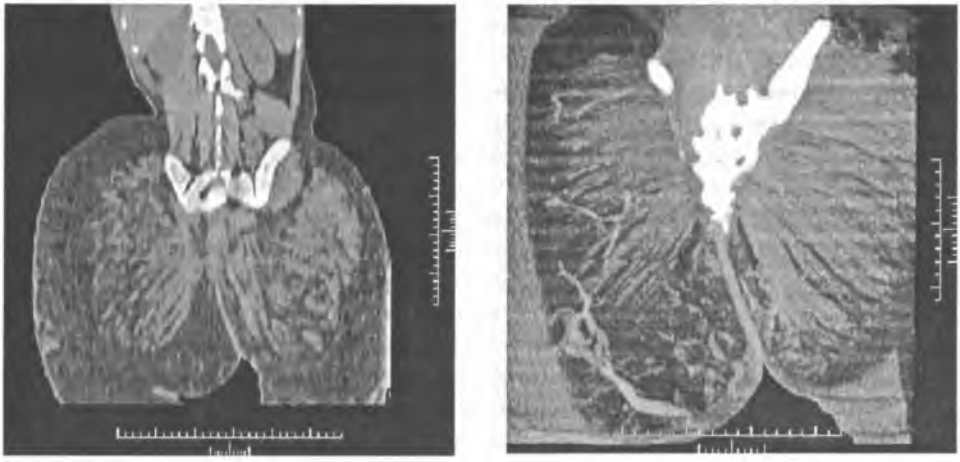

Fig. 3. Symptoms of unilateral hypertrophy of soft tissues (a) with venous malformations (b) typical of KTS

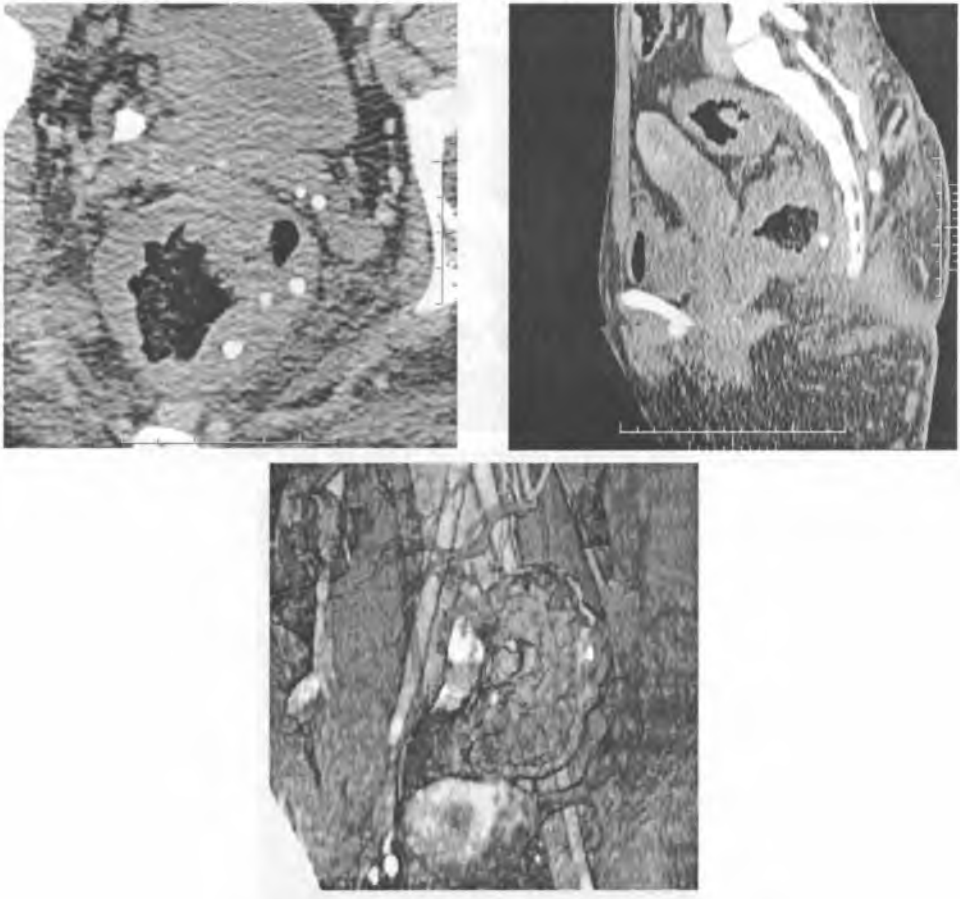

Fig. 4. Circular thickening of the rectal and sigmoid wall with phleboliths typical of hemangioma situated also in the venous plexus of the pelvis; a) oblique MPR view, b) sagittal view, c) 3D volume rendering view

DISCUSSION

Hematuria is a rare presentation in adults with KTS (4, 11). It is usually related to urinary tract hemangiomas, which occur in 3 to 6% of those patients (3). Surgical treatment is required in patients with massive recurrent bleeding (3), in episodic manifestations conservative endoscopic and arteriographic control is suggested (11). In the presented case no typical venous malformations with phleboliths of the urinary tract were identified, however, cystoscopic evaluation of the right ureter suggested the bleeding site location in the right upper urinary tract. The cause of bleeding in the right ureter orifice cannot be definitely determined but in the pathogenesis of the lesions concomitant consumption coagulopathy should be taken into consideration; this kind of a clinical picture is defined as Kasabach-Meritt syndrome (5). An atypical lesion in the central lower pole of the right kidney requires further diagnostic procedures, yet the patient has not been seen to date. Furthermore, the cause of portal hypertension is unclear – in some patients with KTS hypertension may be related to right ventricle failure due to big shunt in microfistulas developing during the disease and chronic coagulopathy.

The characteristic histological features of diffuse cavernous hemangioma of the rectum include: large, thin-walled vascular spaces with clots and scant stroma consisting of fibrous elements and smooth muscles and relatively numerous phleboliths (5, 10). Its invasive character with possible progression to the surrounding structures differentiates the diffuse cavernous hemangioma of the rectum from other vascular lesions of the intestines (1, 2, 10).

Computed tomography in the patient showed multiple KTS-related abnormalities, including the presence of colorectal hemangioma. Unlike other types of examination, CT provides a simple, non-invasive diagnosis of these lesions (8, 10, 12). The application of CT allows not only to diagnose hemangioma, but it is also helpful in assessing trans- and extraparietal extension and range of changes in the colon, important for surgical treatment planning (2). The presence of phleboliths within the thickened colon wall is considered pathognomonic for colorectal hemangioma (1, 5, 8, 10). The intravenous contrast application usually does not cause substantial enhancement of the tumour mass because of low enhancement of vessels caused by slow blood flow accompanied by intrahemangiomatic coagulation (9). Angiography, however, may be valuable in assessing active bleeding sites (10).

Endoscopic evaluation of colorectal hemangioma is limited to its proximal localization, and biopsy of hemangioma may cause life-threatening hemorrhagia (5, 7) as well as endoscopy itself. In rectal localization endoscopic ultrasonography may be useful in estimating the rectum wall status. MRI, particularly with the endorectal coil, is thought to be the examination of choice in the diagnosis of colorectal hemangioma (3, 6). In our country, the availability of MRI is still too limited and the majority of patients are diagnosed with US and CT examinations. Moreover, our patient was referred to angioCT due to hematuria and the intestinal lesion was detected incidentally. However, considering the indefinite picture of the lesion in the right kidney and indications for additional evaluation of the tumour in the large intestine, MRI will be carried out during the next patient's visit.

The use of MSCT provided complex imaging of the abdominal organs and pelvis as well as abnormal vascular structures and lesions of the soft tissues in the iliofemoral region allowing us to assess the extent of lesions in such a big area by means of the multi-phase technique. However, we believe that due to difficulties in precise determination of the course and junctions of the individual vessels, the examinations of choice for evaluating venous vascular malformations of extremities are still phlebography and colour Doppler USG.

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SUMMARY

The paper reports a rare case of an 18-year-old woman with Klippel-Trenaunay syndrome, whose multi-phase multi-slice CT revealed diffuse colorectal hemangioma. The patient presented symptoms of consumption coagulopathy (Kasabach-Meritt syndrome) and intense collateral circulation in the course of portal hypertension. The CT lesions and possibilities of multiformat and 3D volume rendering reconstructions are presented. To our knowledge, this is the first report of diffuse hemangioma of the colon and rectum in a patient with KTS detected with multi-slice CT.

Naczyniak odbytnicy rozpoznany w wielorzędowej tomografii komputerowej u 18-letniej pacjentki z zespołem Klippel-Trenaunaya

W pracy przedstawiono rzadki przypadek 18-letnie chorej z zespołem Klippel-Trenaunaya, u której w wielorzędowej tomografii komputerowej wykryto naczyniaka odbytnicy i esicy. Pacjentka miała objawy koagulopatii ze zużycia (zespół Kasabacha- Meritta) oraz objawy nasilonego krążenia obocznego w przebiegu nadciśnienia wrotnego. Przedstawiono obraz zmian w CT oraz możliwości rekonstrukcji wielopłaszczyznowych i trójwymiarowych. Według naszej wiedzy jest to pierwszy przypadek rozpoznania naczyniaka odbytnicy metodą wielorzędowej tomografii komputerowej u pacjenta z zespołem Klippel-Trenaunaya.